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Intrapericardial Teratoma in an Adult: A Rare Presentation

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Background. Intrapericardial teratomas are rare and usually present early in infancy or childhood.

Procedure. We describe herein a rare case of an adult patient with an intrapericardial teratoma who presented with fever, cardiac arrhythmias, and oppressive substernal chest pain. Preoperative diagnosis was suggested by echocardiography and computerized tomography of the chest. The tumor weighed 530 g and its histologic features were those of a mature cystic teratoma. It was excised totally and 10 years' follow-up revealed no evidence of residual disease.

Discussion. Our patient is one of the very few adult patients with intrapericardial teratomas who was treated successfully with surgery. Both echocardiography and tomography of the chest suggested the diagnosis and delineated the relationship of the tumor to the great vessels.

Conclusion. The diagnosis of Intrapericardial teratomas is suspected by echocardiography and/or tomography of the chest and confirmed by specific histologic features. These tumors should be excised whenever detected. Med. Pediatr. Oncol. 30:249–251, 1998.

Key words: intrapericardial teratoma; adult; echocardiography; computerized tomography of the chest; histologic features; excision

INTRODUCTION

Teratomas are tumors of embryonic origin composed of elements derived from all three germ cell layers in varying degrees. In the anterior mediastinum, they arise from remnants of the third branchial pouch in close proximity to the thymus gland. When located in the posterior mediastinum, in 3–8% of all mediastinal teratomas [1], it is postulated that they arise from notochondral remnants.

Intrapericardial teratomas are rare tumors with only about 60 cases reported in the literature, and the majority of the patients are of the pediatric age group [2]. We report a rare case of pericardial teratoma in a 26-year-old, that was totally excised with no recurrence on 10 years' follow-up.

CASE REPORT

J.G. is a 26-year-old male who was doing well until 2 months prior to his presentation to our hospital when he felt an acute oppressive chest pain accompanied with fever. He was admitted to a local hospital where a chest X-ray film revealed an enlarged cardiac silhouette. He was started on intravenous antibiotics and defervesced 5 days later. He also developed atrial flutter and was treated medically and discharged home. Two weeks later, the fever recurred and the patient was admitted to our hospital where an echocardiogram revealed a large multiloculated cystic mass impinging on the right atrium and compressing the right ventricular outflow tract. A computerized tomography of the chest showed a large intrapericardial multicystic and lobulated ovoid mass adherent to the aorta and displacing the right atrium (Fig. 1). The diagnosis of intrapericardial teratoma was entertained and the patient was explored through a median sternotomy. The tumor was all intrapericardial. It was shelled from the inner pericardial surface, right atrium, and right ventricle. The tumor was adherent to the aortic adventitia at the root of the ascending aorta, which necessitated excision with the tumor in toto.

The multilobulated mass measured 19 × 10 × 9 cm and weighed 530 g. Its cut surface disclosed multicystic cavities filled with mucoid material and focal areas of calcification and hemorrhage. The tissue was fixed in 10% buffered formalin. Paraffin-embedded sections were stained with hematoxylin and eosin. Microscopically, the mass was composed of a heterogeneous population of mature cells representing all three germ layers. The endodermal component mainly consisted of large spaces lined by either ciliated respiratory or mucus-secreting gastrointestinal epithelium. The ectodermal component was made up of scattered patches of glial tissue often containing microcalcifications, and spaces lined by a

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Fig. 1. CT scan section through the mid right atrium showing the intrapericardial mass impinging but separate from the right atrium. L = left; R = right.

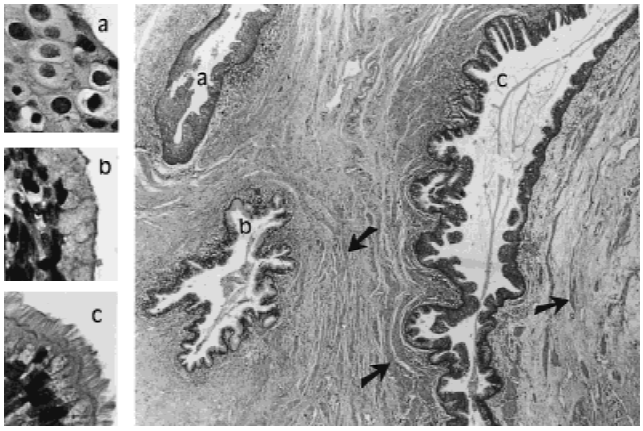

Fig. 2. **Right:** A section of the tumor showing three spaces, each lined by a different type of mature epithelium. These are surrounded by a fibrous stroma containing strands of smooth muscle (arrows) (H & E, ×50). Higher magnification of the epithelial linings are seen in the insets: squamous **(a),** mucus-secreting **(b),** and ciliated **(c)** (H & E, × 200).

nonkeratinized squamous epithelium. Mesoderm was represented by a fibroadipose tissue stroma containing strands of smooth muscle and occasional islands of hyaline cartilage (Figs. 2–4). The histologic features were those of a mature cystic teratoma. The patient did well and 10 years' follow-up revealed no evidence of residual disease.

DISCUSSION

Mediastinal teratomas may occur at any age, but are more frequently seen in adults aged from 20 to 40 years [3]. In infants, teratomas are the most common anterior mediastinal neoplasms, whereas in adults, the anterior

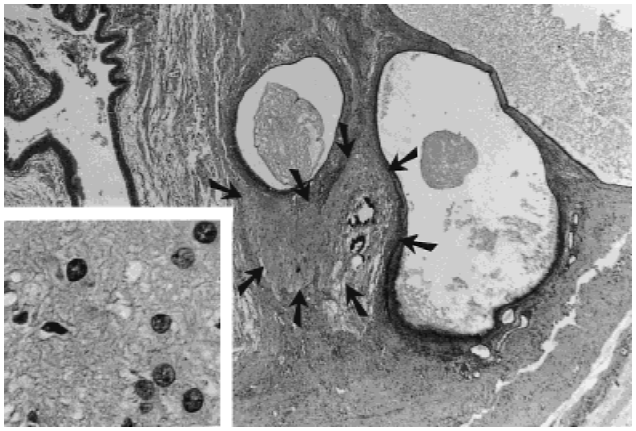

Fig. 3. A focus of mature brain tissue with microcalcifications (arrows) between two cystic spaces (H & E, ×50). The inset demonstrates the glial nature of this tissue (H & E, ×200).

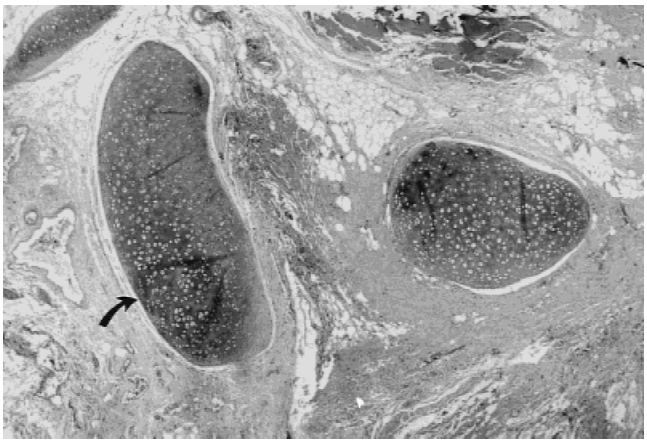

Fig. 4. Fibroadipose tissue stroma with an arrow showing one of three islands of cartilage (H & E, ×80).

mediastinum, second to the ovary and testicles, is the commonest site for teratomas. Teratomas within the pericardium are the least common, most cases occurring in infancy and childhood. The tumor has no sex predilection [2] and may present in a variety of ways. In infancy, low cardiac output state and cyanosis usually develop; however, at puberty, presentation is more benign in the form of chest discomfort, pulmonary infection due to lung compression, or secondary bacterial infection of the tumor itself [1]. Arrhythmias are not uncommon.

In the rare cases where teratomas are intrapericardial, they tend to present early in infancy and childhood because of symptoms related to compression of the heart and great vessels. The majority of the cases reported in the literature and especially those that were successfully treated with surgery belonged to the pediatric age group. Our patient is one of the very few adult patients with intrapericardial teratomas who was treated successfully

with surgery. Our patient presented with atrial flutter indicating a possible compression of the right atrium by the tumor. Both echocardiography and tomography of the chest suggested the diagnosis and delineated the relationship of the tumor to the great vessels. Fever was presumed to be secondary to tumor infection. Calcifications are rarely detected in intrapericardial teratoma, unlike teratoma of the posterior mediastinum.

Intrapericardial teratomas should be excised whenever detected because they carry a risk for malignant transformation [4]. A second reason is that their malignant behavior, in proximity to such a vital organ system, may lead to compression and arrhythmias, and, third, they carry the likelihood of becoming infected. Our patient had complete resection of the tumor, and is asymptomatic 10 years later, with no apparent residual disease.

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